PIR QUIZ

9. In the immediate neonatal period, the *least likely* cause of status epilepticus is:
   A. Hemorrhage into the central nervous system.
   B. Hypoxic-ischemic encephalopathy.
   C. Inborn errors of metabolism.
   D. Infection.
   E. Unsuspected parental abuse.

10. In early childhood, the *most likely* cause of status epilepticus is:
   A. Chromosomal disease with central nervous system abnormalities.
   B. Drug overdose.
   C. Febrile seizure lasting longer than 30 minutes.
   D. Metabolic disease with lactic acidosis.
   E. Unsuspected head trauma.

11. The *most correct* statement regarding absence status epilepticus is that:
   A. Abnormalities in the electroencephalogram require hyperventilation for detection.
   B. It occurs frequently in those younger than 6 years of age.
   C. It often is the first sign or symptom of an intracranial neoplasm.
   D. It typically manifests as a drowsy, confused state in a patient who has had prior seizures.

12. The recommended *initial* pharmacologic approach to the treatment of status epilepticus is:
   A. Lorazepam 0.05 to 0.1 mg/kg intravenously.
   B. Nitrous oxide by inhalation.
   C. Pentobarbital 3 to 5 mg/kg intravenously.
   D. Phenytoin 15 to 20 mg/kg intramuscularly.
   E. Sodium valproate syrup 20 mg/kg in water rectally.

13. The *most urgent* laboratory test(s) to perform in a patient who has status epilepticus is:
   A. Blood glucose by Dextrostix®.
   B. Blood pH and lactic acid levels.
   C. Complete blood count.
   D. Serum calcium and magnesium levels.
   E. Urine toxicology screen.

The Diagnosis of Rheumatic Fever

Acute Rheumatic Fever. Wald E. *Curr Probl Pediatr.* 1993;23:264–270

Rheumatic Fever: Keeping up with the Jones Criteria. Forster J. *Contemp Pediatr.* 1993;10:51–60

Treatment of Acute Streptococcal Pharyngitis and Prevention of Rheumatic Fever: A Statement for Health Professionals. Dajani A, Taubert K, Ferrieri P, Peter G, Shulman S, and American Academy of Pediatrics Committee on Infectious Diseases and the American Heart Association Committee on Rheumatic Fever, Endocarditis, and Kawasaki Disease of the Council on Cardiovascular Disease in the Young. *Pediatrics.* 1995;96:758–764

Acute rheumatic fever (ARF) was recognized initially in the late 19th century and followed a declining pattern of incidence in the United States until the mid-1980s. It remains one of the primary causes of acquired heart disease worldwide. A resurgence of ARF since 1984 prompted the medical community to review the early signs and symptoms of an illness that was considered to be uncommon. Traditionally, ARF was thought to be a disease of the inner-city poor and military recruits, but in recent resurgences, rural and suburban communities have been affected as well.

The most common clinical manifestations of ARF in recent outbreaks in the United States were arthritis and carditis. During these outbreaks the majority of patients showed one major manifestation, but two major manifestations (carditis and arthritis or carditis and chorea) also were seen frequently. Many of the patients diagnosed as having ARF during these epidemics had no recognizable prodrome that would have brought them to medical attention. A history of symptomatic pharyngitis often was absent. It is important to remember that the throat culture frequently is negative by the time rheumatic fever develops. These facts emphasize the need to consider ARF in the appropriate clinical setting and use the streptococcal enzyme tests to establish a diagnosis.

Cardiac involvement often is established by the finding of a new murmur of mitral or aortic insufficiency. Pancarditis, with pericardial and myocardial involvement, can be seen along with valvulitis. Mitral regurgitation, heard best at the apex, is generally of moderate-to-high intensity throughout systole. Aortic insufficiency is a basal diastolic murmur that is usually high-pitched and blowing and decreases in intensity toward the end of diastole. Currently, echocardiography is used to confirm the auscultatory findings, but hemodynamically insignificant echocardiographic findings alone are not considered sufficient to diagnose carditis.

The classic migratory polyarthritis of ARF often involves the extremities (elbows, wrists, knees, and ankles) and is extremely painful. It usually presents early in the disease and is short-lived (<4 weeks). It is exquisitely responsive to standard anti-inflammatory therapy. Symptoms of chorea present late (unlike arthritis or carditis), usually months after the initial pharyngitis. The process is self-limiting and reversible.

The Jones Criteria for the diagnosis of ARF, published originally in 1944, have been updated several times, most recently in 1992. The 1992 update differs from prior versions in its strong focus on identifying acute episodes of rheumatic fever. Whereas previously two major or one major and two minor criteria were required to fulfill the diagnostic profile, evidence of a preceding streptococcal infection (such as an elevated antisteptolysin O [ASO] titer) *in addition* to two major or one major and two minor manifestations now are needed for diagnosis (Table). It is important to note that the Jones Criteria are not all-inclusive. For example, carditis or especially chorea can be the sole presenting symptom.

The overall incidence of streptococcal pharyngitis has remained essentially unchanged during this century. The underlying reasons for the decrease in ARF during this time has been attributed to the possibility that some M types are more "rheumatogenic" than others. Rheumatogenicity may be due to the presence of an M-associated protein I surface antigen and the absence of a serum opacity reaction (SOR) in these

M types. The concept of rheumatogenicity is particularly attractive because other explanations, including improved hygiene, standards of living, and the availability of antimicrobial treatment, cannot account for the previous decline in disease. Group A streptococcal serotypes are known to increase and decrease in different geographic locations. A resurgence of ARF could be attributed to the introduction of a rheumatogenic strain in a particular geographic area. Host susceptibility, including predisposing genetic factors, also may influence the likelihood of developing ARF after a streptococcal pharyngeal infection.

The diagnosis of ARF is based on the finding of recent streptococcal infection and clinical findings consistent with the major and minor criteria. Laboratory evaluation of ARF must focus primarily on the identification of antecedent group A streptococcal infection. A positive throat culture or rapid antigen test confirms a recent infection. Rapid antigen tests are used very frequently as screening tests for group A streptococcal infection, but throat culture remains the definitive test to establish the diagnosis. When the throat culture is negative, serologic tests, such as elevated ASO, antideoxyribonuclease B (anti-DNase B), or antihyaluronidase titers are used. The ASO titer is the serologic test used most commonly, but there are no established criteria for the degree of elevation of the ASO that correlates with a definitive diagnosis. In some patients, the ASO titer may be elevated only moderately. If the ASO titer is normal and streptococcal infection is suspected, one of the other serologic tests can be used as confirmatory evidence of recent infection. Using two or three enzyme tests improves the recognition of infection.

TABLE. The 1992 Jones Criteria

1. Evidence of a preceding group A streptococcal infection:
   Elevated or rising ASO titer or
   Positive throat culture or
   Positive rapid antigen test.

Plus

2. Either **two major** or **one major and two minor** manifestations:
   Major manifestations: Carditis, polyarthritis, Sydenham chorea, erythema marginatum, subcutaneous nodules.
   Minor manifestations: Arthralgia, fever, elevated acute-phase reactants (ESR, C-reactive protein), prolonged PR interval.

Recommendations for the primary prevention of rheumatic fever (treatment of streptococcal pharyngitis) still include intramuscular penicillin G and oral penicillin V or erythromycin estolate/ethylsuccinate. Other alternatives include azithromycin and the oral cephalosporins (see Pichichero in this issue of *Pediatrics in Review*). It is essential to employ continuous antibiotic prophylaxis to prevent recurrences of rheumatic fever due to subsequent streptococcal infection. Benzathine penicillin G 1.2 million units intramuscularly remains the treatment of choice for prophylaxis and is administered every 3 or 4 weeks. Alternative regimens for prophylaxis include daily penicillin V, sulfadiazine, or erythromycin. There is recent evidence that for patients at particularly high risk, such as those living in endemic areas or those who have chronic rheumatic heart disease, an every 3-week regimen of intramuscular penicillin prophylaxis is more protective. This prophylactic regimen does not substitute for the standard bacterial endocarditis prophylaxis required for patients who have rheumatic heart disease. The duration of prophylaxis for patients who do not have carditis is 5 years or until age 21 (whichever is longer). For patients who have carditis but no residual heart disease, prophylaxis is continued for 10 years or well into adulthood (whichever is longer). Those patients who have residual heart disease from carditis are treated at least until age 40 or may receive lifelong prophylaxis.

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